**Imaging features of pulmonary sarcoidosis - a pictorial essay**

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Learning objectives

- To review the basic aspects of pulmonary sarcoidosis;

- To present usual and unusual patterns of thoracic sarcoidosis, as seen on plain film and CT.

Background

DEFINITION AND EPIDEMIOLOGY

Sarcoidosis is a multiorgan granulomatous disease of unknown cause. It has a worldwide distribution and typically affects young to middle-aged adults, with a slightly higher incidence in women.

CLINICAL PRESENTATION

Sarcoidosis involves the thorax in about 90% of patients and lung involvement accounts for most of the morbidity and much of the mortality associated.

Clinical presentation is variable. Approximately 50% of patients are asymptomatic. The remainder present with either respiratory symptoms (e.g. cough and dyspnoea) or skin changes (e.g. erythema nodosum, lupus pernio, scars, plaques).

Löfgren syndrome is an uncommon but specific acute presentation with a better prognosis.

Respiratory function tests tend to demonstrate a restrictive pattern with reduced lung volumes and reduced diffusion capacity. Obstructive components usually seen in patients who also smoke, and are largely attributable to the later.

IMAGING FEATURES
Thoracic sarcoidosis has been called "the great mimic"; it manifests with various patterns at radiologic imaging, necessitating an initially broad differential diagnosis that includes lymphoma, tuberculosis, and many other causes of chronic pulmonary infiltrates.

In typical cases, chest radiography may be sufficient to establish the diagnosis with little margin of error, with bilateral hilar lymph node enlargement being the most common finding, followed by interstitial lung disease, but there are many possible presentations.

CT plays a critical role in atypical and complicated cases and is more sensitive for the detection of adenopathy and subtle parenchymal disease.

**STAGING AND PROGNOSIS**

Several years ago, Siltzbach developed a sarcoidosis staging system based on the pattern of chest radiographic findings, a system still widely used because of its great prognostic value.

It defines the following five stages of sarcoidosis:

- **stage 0**: normal appearance at chest radiography;
- **stage I**: lymphadenopathy only;
- **stage II**: lymphadenopathy and parenchymal lung disease;
- **stage III**: parenchymal lung disease only;
- **stage IV**: pulmonary fibrosis.

In most patients the condition either regresses or remains stable. Progression to pulmonary fibrosis occurs in approximately one-fourth of patients. Generally, pulmonary function worsens with more advanced disease stages, but the radiographic stage does not correlate well with the severity of pulmonary function abnormalities.

Spontaneous remission occurs in 60%-90% of patients with stage 1 disease, in 40%-70% with stage 2 disease, in 10%-20% with stage 3 disease, and in 0% with stage 4 disease.3

**Imaging findings OR Procedure details**

**PLAIN FILM**
Typical plain film findings include mediastinal and bilateral hilar lymphadenopathy, parenchymal opacities, and, in more advanced cases, parenchymal fibrosis.

As mentioned in the background section, pulmonary sarcoidosis may be classified on a chest x-ray into 5 stages:

- **stage 0**: normal chest radiograph
  - 5 - 10% of patients at presentation
- **stage I (Figure 1)**: hilar or mediastinal nodal enlargement only
  - 45 - 65% of patients at presentation
  - 60% go onto complete resolution
- **stage II (Figure 2)**: nodal enlargement and parenchymal disease
  - 25 - 30% of patients at presentation
- **stage III (Figure 3)**: parenchymal disease only
  - 15% of patients at presentation
- **stage IV (Figure 4)**: end-stage lung (pulmonary fibrosis)

Besides these classical presentations, there are atypical findings that can appear in the X-Ray, including:

- Calcified lymphadenopathy (Figure 5);
- Pleural effusion (Figure 6);
- Pleural thickening and pneumothorax (Figure 7);
- Alveolar opacities (Figure 8);
- Multiple nodules mimicking metastases (Figure 9);
- Miliary sarcoidosis (Figure 10).

**CT/HRCT**

At CT it often presents as subcarinal and symmetrical hilar lymph node enlargement (Figure 11) with the typical perilymphatic distribution of nodules (Figure 12).

The nodules predominate in the mid to upper lung zones and may coalesce or clump into larger masses (Figure 13).

At end-stage disease, it may present as fibrotic conglomerate masses causing traction bronchiectasis in the central and upper lung (Figure 14).
High-resolution CT (HRCT) has proved superior to conventional CT for assessing subtle parenchymal details and discriminating between inflammation and fibrosis in patients with pulmonary sarcoidosis. Fibrosis signs at CT/HRCT include the following (Figures 14 and 15):

- posterior displacement of upper lobe and main bronchi (usually the earliest HRCT finding);
- architectural distortion;
- hilar retraction;
- upper lobe volume loss;
- broad and coarse septal bands;
- honeycombing;
- large bullae.

Uncommon or atypical features include, besides those mentioned above regarding plain film findings and depicted at CT/HRCT in Figures 16 to 18, the following:

- Unusual lymphadenopathy distribution;
- Cavitation;
- Aspergillomas;
- Irregular thickening of interlobular septa and bronchovascular bundles;
- Asymmetrical and predominantly basal lung involvement;
- Unilateral sarcoidosis;
- Airway involvement;
- Vascular involvement.

Images for this section:
Fig. 1: Stage I Sarcoidosis: PA Chest X-Ray shows bilateral lobulated bulging of the pulmonary hila (red arrows).
Fig. 2: Stage II Sarcoidosis: PA Chest X-Ray shows left hilar node enlargement (red arrow) and multiple bilateral millimetric nodules in the parenchyma (green arrow), confirmed by CT.
Fig. 3: Stage III Sarcoidosis: PA Chest X-Ray shows a few ill-defined small nodular opacities located in the upper third of the right lung (green arrow), as well as some suggestion of reticular septal thickening (red arrow) in the middle third of the same lung.
Fig. 4: Stage IV Sarcoidosis: PA Chest X-Ray shows multiple irregular reticular opacities irradiating from the hila due to fibrosis.
**Fig. 5:** Chest X-Ray showing calcified lymph nodes in the right paratracheal (red arrow) and hilar compartments (green arrows). On the right side image, we can see calcified supraclavicular nodes (arrows).
Fig. 6: Chest X-Ray shows right-sided pleural effusion (red arrow) and pneumothorax (green arrow), with tenting of the hemidiaphragms.
Fig. 7: Chest X-Ray shows evidence of pleural thickening (red arrows) and subtle pneumothorax (green arrow). There is also bilateral upper lobe fibrosis with hilar retraction.
**Fig. 8:** PA Chest X-Ray depicting multiple ill-defined nodular opacities in the middle third of both lungs, which coalesce centrally assuming the form of a large air-space consolidation with air bronchograms.
Fig. 9: Chest X-Ray showing multiple and bilateral pericentimetric nodules, mimicking metastatic disease.
Fig. 10: Chest X-Ray shows multiple, diffuse, milimetric nodules in both lungs.
**Fig. 11:** Chest CT shows bilateral hilar (green arrows) and subcarinal lymphadenopathy (red arrow).
**Fig. 12:** HRCT showing a typical micronodule distribution along the bronchovascular bundles (green arrow) and subpleural region (red arrow).
Fig. 13: HRCT shows multiple micronodules in the middle zone of both lungs coalescing centrally.
Fig. 14: HRCT shows architectural distortion of the right upper lobe, intra and interlobular septal thickening (a) and traction bronchiectasis.
**Fig. 15:** HRCT coronal reformation showing extensive signs of fibrosis with upper lobe predominance.
Fig. 16: Chest CT depicting calcified lymphadenopathies (green arrows).
**Fig. 17:** HRCT coronal reformation showing multiple irregular, ill-defined nodules in both lungs - alveolar sarcoidosis.
Fig. 18: HRCT depicting multiple peri-centimetric nodules in both lungs (arrows) - sarcoidosis mimicking metastatic disease.
Conclusion

In most patients with sarcoidosis, pulmonary manifestations become evident at some point. However, it is important to recall that imaging features are nonspecific or atypical in 25%-30% of patients, and in another 5%-10%, no abnormalities are seen at thoracic imaging.

CT is better than plain radiography in detecting pulmonary findings. HRCT goes even further in depicting parenchymal detail.

It is essential for the radiologist to recognize both typical and atypical radiologic manifestations in these techniques.

References


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